

Cobb's Syndrome : A Case Report¹

Eun Ja Lee, M.D., Si Won Kang, M.D., Kyung Sub Shinn, M.D.

Cobb's syndrome is characterized by spinal and vertebral hemangiomas with a skin nevus at the same metameric level. We report a case of Cobb's syndrome in a young female patient with radiologic findings including simple X-ray, magnetic resonance imaging, and angiography.

Index Words : Spine, angiography
Spine, MR
Spine, neoplasms

Cobb's syndrome consists of vascular skin nevi associated with an angioma in the spinal cord. This condition is a rare clinical entity and only 28 cases are documented in the literature (1, 2). It must be differentiated from other syndrome characterized by vascular cutaneous lesion associated with central nervous abnormalities such as Sturge-Weber, Osler-Weber-Render, Fabry-Anderson, von Hippel-Lindau, and ataxia telangiectasia (1).

Case Report

A 14-year-old girl was admitted for progressive weakness of the lower extremities, port-wine nevus on the back and right anterior chest (present from birth), and multiple skin nodules on the trunk. When she was five, a 2cm cutaneous nodule, an angioma, was noted on the back and excised. Since then, multiple skin nodules developed and progressively increased in size and number. For a year she had suffered from progressive weakness of the lower extremities and physical therapy resulted in no improvement of symptoms. Neurologically, cranial nerve examination was normal. Weakness of the right lower extremity was grade four positive (IV⁺), and of the left, grade four zero (IV⁰).

Hypoesthesia below dermatome level T3 was noted and knee jerk was hyperactive. Babinski's sign was bilaterally positive. Anal sphincter tone was decreased and there was chronic constipation.

A plain radiograph of the spine showed coarse vertical striations in vertebrae from C7 to L5, suggesting typical vertebral hemangioma. Spinal MRI revealed a long band-like posterior extradural mass from C7 to L5 with mixed low and high signal intensities (predominantly low signal intensity) on sagittal T1-weighted images, due to subacute and chronic hemorrhage (Fig. 1A). On sagittal T2-weighted images, the mass showed predominantly high signal intensity intermingled with dot-like low signal intensities (Fig. 1B), it was diffusely and intensely enhanced after gadolinium-enhancement (Fig. 1C, D). Internal low signal dots seen on T2-weighted images were poorly enhanced. The lesion appeared to be extended into intervertebral foramina in association with foraminal widening (Fig. 1D); the dural sac was displaced anteriorly by the mass.

Abnormal signal changes were also noted in vertebrae including bodies and neural arches from C7 to L5. Vertebral bodies showed low signal intensity on T1-weighted image, and high signal intensity on T2-weighted image, replacing normal fatty marrow. Patchy high signal intensity areas on T1- and T2-weighted images were present, however, in several vertebral bodies (Fig. 1A, B). Involved neural arches were hypertrophied. Postcontrast images showed intense enhancement of vertebrae. There was no definite evidence of involvement of the spinal cord, and brain

¹Department of Radiology, Taejon St. Mary's Hospital, The Catholic University of Korea

Received September 4, 1996; Accepted October 23, 1996

Address reprint requests to: Eun Ja Lee, M.D., Department of Radiology, Taejon St. Mary's Hospital, The Catholic University of Korea, #520-2 Taehung-dong Taejon, 301-012, Korea.

Tel. 82-42-220-9625 Fax. 82-42-252-6807

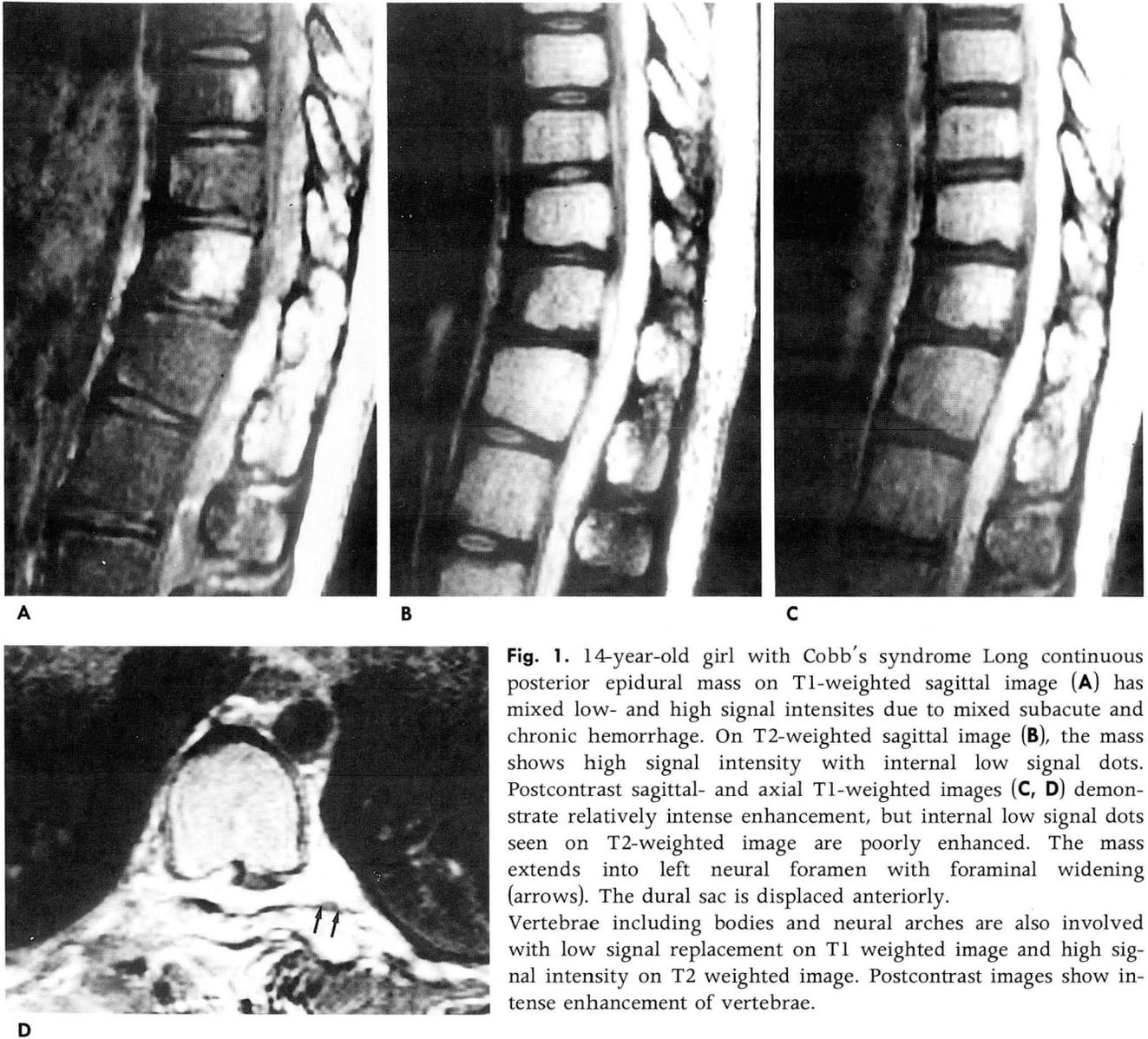


Fig. 1. 14-year-old girl with Cobb's syndrome Long continuous posterior epidural mass on T1-weighted sagittal image (A) has mixed low- and high signal intensities due to mixed subacute and chronic hemorrhage. On T2-weighted sagittal image (B), the mass shows high signal intensity with internal low signal dots. Postcontrast sagittal- and axial T1-weighted images (C, D) demonstrate relatively intense enhancement, but internal low signal dots seen on T2-weighted image are poorly enhanced. The mass extends into left neural foramen with foraminal widening (arrows). The dural sac is displaced anteriorly. Vertebrae including bodies and neural arches are also involved with low signal replacement on T1 weighted image and high signal intensity on T2 weighted image. Postcontrast images show intense enhancement of vertebrae.

MRI revealed no definite abnormality. Spinal angiography with selection of the right side of T3, 4 and 6, the left side of the 8th, 9th and 12th intercostal arteries, and the left L1 lumbar artery showed increased vascularity of epidural and cutaneous hemangioma, and the dense stain of a vertebral hemangioma supplied by the same intercostal artery (Fig. 2A, B).

A total laminectomy from T11 to L1 level was performed. At operation, the mass in the posterior epidural space was pinkish, rubbery and easily removed and the bone was hypertrophied and osteoporotic. The spinal canal was narrowed due to hypertrophied bone, and there was severe bone bleeding.

On histopathologic examination, vertebral and epidural masses were found to be cavernous hemangiomas, and excised cutaneous nodules were capil-

lary hemangiomas.

Discussion

Cobb's syndrome is a segmental cutaneous hemangioma of the thorax combined with a hemangioma of the spinal meninges in corresponding segments (3-7). Two elements are necessary to justify a diagnosis of cutaneomeningospinal angiomas, as described by Cobb: (a) a vascular skin nevus and (b) an intraspinal angioma(3).

In 1929, Globus and Doshay (7) reviewed intraspinal vascular anomalies of the spinal cord in Cobb's syndrome, as follows: (a) dilatation of spinal veins (also called pial hemorrhoids and angioma venosum racemosum), (b) arterial or arteriovenous aneurysms of

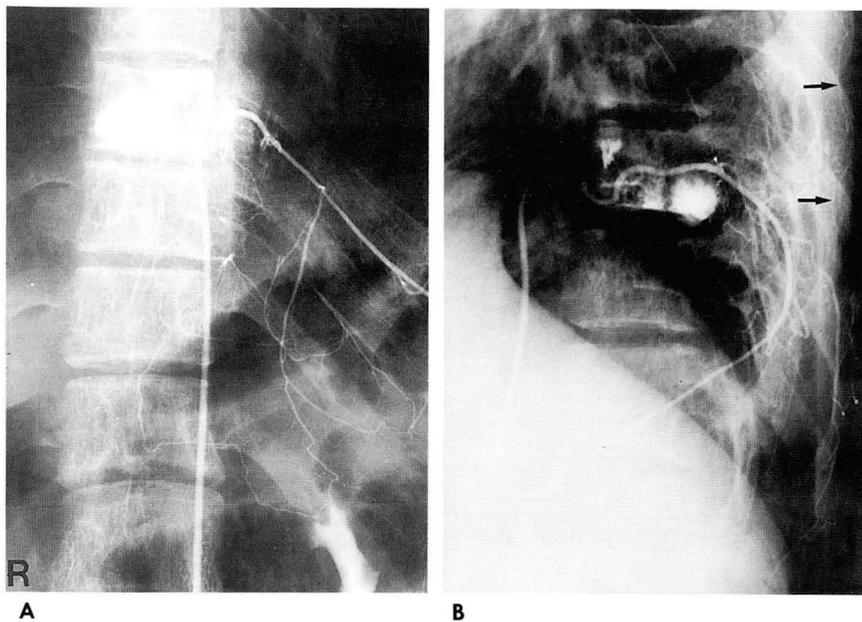


Fig. 2. Spinal angiograms (A, B) of left T9 intercostal artery show increased vascularity of intraspinal and cutaneous (arrows) hemangiomas and dense stain of vertebral hemangioma supplied by same intercostal artery.

the spinal vessels, (c) hemangiomas; ① intramedullary hemangiomas; ② extramedullary pial hemangiomas; ③ epidural hemangiomas; ④ vertebral hemangiomas (7). The case presently under review showed epidural and vertebral hemangioma involving levels C7 to L5 associated with cutaneous hemangioma of the same dermatomes; there was, however, no definite evidence of involvement of the spinal cord. From an embryological point of view, the occurrence of these metameric cutaneous, vertebral and spinal cord vascular malformation may relate to the common origin of the blood supply to the vertebrae and the spinal cord from the segmental dorsolateral arteries (2) (Fig. 3).

In Cobb's syndrome, males outnumbered females and neurologic problems usually occurred during childhood or adolescence. No inciting event except exercise has been reported and familial cases of Cobb's syndrome have not been reported (1). The neurologic features of Cobb's syndrome were the clinical expression of intraspinal angioma. The frequent initial symptoms were pain or motor disorders (paraplegia or monoplegia, recurrent or progressive). Sphincter upset, paraesthesiae, and hypoesthesiae are, in addition, less common (2, 3). The results of a lumbar puncture were usually normal, but occasionally there would be evidence of spinal blockage, hemorrhage, and increased protein and cell counts.

Selective arteriography demonstrated opacification of both the spinal and cutaneous vascular malformation from the same intercostal or lumbar artery, confirming the similar metameric origin of both lesions; selective spinal angiography might aid not only diag-

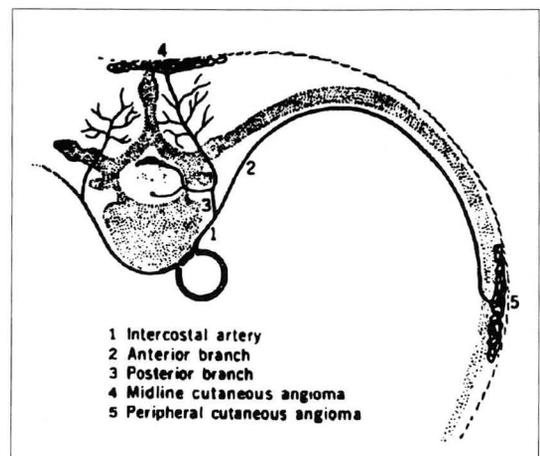


Fig. 3. Diagrammatic cross-section showing the common origin of the blood supply to cutaneous and spinal-cord angiomas.

nosis but also treatment. Miyatake et al (2) reported that a Cobb's syndrome patient with extradural vascular malformation (such as arteriovenous fistula or varix) was treated with liquid embolization using ethylene vinyl alcohol (2).

Although invasive examinations such as spinal angiography have been used in the diagnosis of spinal vascular malformation, noninvasive MR imaging detect both vertebral angiomas and spinal vascular malformation. Patients with motor or sensory disturbances accompanied by a metameric nevus should therefore be screened with MR imaging to disclose these spinal vascular malformations.

References

1. Jessen RT, Thompson S, Smith EB. Cobb syndrome. *Arch Dermatol* 1977; 113: 1587-1590
2. Miyatake SI, Kikuchi H, Koide T, et al. Cobb syndrome and its treatment with embolization: Case report. *J Neurosurg* 1990; 72: 497-499
3. Kissel P, Dureux JB. *Cobb syndrome: Cutaneomeningospinal angiomas*. In Bruyn GW, Vinken PJ, eds. *Handbook of clinical Neurology*. vol 14. New York, North Holland Publishing Co, 1972: 429-445
4. Short MP, Adams RD. *Neurocutaneous disease*. In: Fitzpatrick TB, Eisen AZ, Wolff K. *Dermatology in general medicine*. 4th ed. Philadelphia: Mcgraw-Hill Inc. 1993: 2249-2253
5. Baraitser P, Shieff C. Cutaneomeningo-spinal angiomas: The syndrome of Cobb. *Neuropediatrics* 1990; 21: 160-161
6. Silverman S. Vascular tumours of the spinal cord associated with skin haemangiomas. *Br J Surg* 1945; 33: 307-311
7. Johnson LM, Texx N. Clinical notes, suggestions and new instruments: Epidural hemangioma with compression of spinal cord. *JAMA* 1938; 110: 119-122

대한방사선의학회지 1997; 36: 33-36

콕씨 증후군: 1예 보고¹

¹대전성모병원 진단방사선과

이 은 자 · 강 시 원 · 신 경 섭

콕씨 증후군은 척추 및 척수의 혈관종을 동반하면서 동일 체절역에 피부 모반을 특징으로 한다. 저자들은 젊은 여자 환자에서 발생한 콕씨 증후군을 단순 X-선촬영, 혈관조영 및 자기공명영상 등의 방사선학적 소견과 함께 보고하고자 한다.